

## Pediatric case of Graham Little Piccardi Lassueur syndrome – A rare entity

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**Summary** Graham Little Piccardi Lassueur syndrome (GLPLS) is a rare dermatosis characterized by patchy cicatricial alopecia of scalp, rapidly developing keratosis pilaris like follicular papules over trunk and extremities, and noncicatricial loss of axillary and pubic hair. This syndrome which is mostly seen in middle aged post-menopausal females (between ages 30-70) has rarely ever been described in the pediatric age group. We report a case of a 15 year old girl presenting to us with this rare syndrome.

**Keywords:** Graham Little Piccardi Lassueur syndrome, alopecia, lichen planopilaris, pediatric

### 1. Introduction

Graham Little Piccardi Lassueur syndrome (GLPLS) was first described by Piccardi (1914) who reported a case with progressive cicatricial scalp alopecia associated with non-cicatricial alopecia of axillary and pubic areas and a follicular lichenoid eruption on the trunk and extremities. In 1915, Ernst Graham-Little also published a similar case observed by Lassueur, which he called 'folliculitis decalvans et atrophicans' (1). GLPLS is a rare variant of lichen planopilaris usually occurring in middle-aged Caucasian women (2-6). No pediatric case has been reported till date. Herein, we present a case of E/beta thalassemia with GLPLS and its classical triad of clinical features.

### 2. Case Report

A 15 year old girl presented with intermittent episodes of mild yellowish discolouration of eyes since 5 years of age, each episode lasting 7-10 days which

spontaneously disappeared without any medications only to recur 2 months later. These episodes happened several times, each of similar duration and symptom free intervals. Her mother noticed poor growth during this period compared to her siblings. Six months before presentation, she noticed multiple discrete small spiky black spots over the skin of epigastric region. The lesions gradually increased over the whole anterior abdominal wall within 2-3 months (Figure 1). There was associated scalp hair loss over the past 6 months with 2 small areas of baldness with distinct margin over left parietal region (Figure 2) and left posterior auricular region. A patchy lesion over medial aspect of right leg with distinct margin had also appeared progressively increasing in size over last 2-3 months. Antenatal and birth history were uneventful. She was developmentally appropriate and had good scholastic performance. She had not attained menarche and there was no development of her secondary sexual characters. She never required any hospitalization or any blood transfusions during this period. Topical steroid ointments were prescribed in the past but did not relieve her symptoms. On general survey, there was mild pallor and icterus. She was stunted and had no hair over the both axilla (Figure 3) and pubic region with no breast development. There were plenty of discrete black spiky elevated spots dispersed over whole anterior abdomen. There was 2 areas of alopecia areata over scalp and a 3 cm by 4 cm patchy lesion with distinct

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**Figure 1. Multiple discrete black spiky elevated lesions over skin of abdomen.**

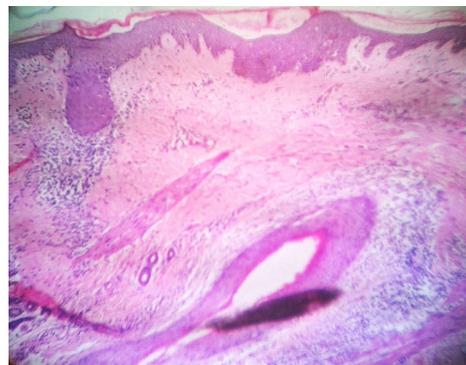


**Figure 2. Cicatricial scalp alopecia over left parietal region.**

margin over the medial aspect of right leg. On systemic examination there was hepatosplenomegaly, both being 2 cm from costal margin, firm in consistency and nontender. Her laboratory reports showed a microcytic hypochromic anemia with haemoglobin 8 g/dL, MCV-62 fl, MCH-18.4 pg, MCHC-24.7 g/dL, red cell distribution width-23.1%. Liver function tests showed total bilirubin was 2.32 mg/dL and conjugated fraction was 1.58 mg/dL with normal transaminases. Thyroid stimulating hormone level was 2.62  $\mu$ IU/mL. Haemoglobin electrophoresis was done in two occasions and showed haemoglobin F was 23-27.3%, haemoglobin A was 6.2-9.9%, haemoglobin A2+E was 49.8-54.8%. Haemoglobin electrophoresis of her mother showed haemoglobin E carrier and father showed beta thalassemia carrier. Our dermatology department diagnosed skin lesions over abdomen as Lichen planopilaris and leg lesions as lichen planus.



**Figure 3. Bilateral loss of axillary hair.**



**Figure 4. Microscopic findings.** Skin biopsy showed perifollicular lymphocytic infiltrate with damage to follicular basal cells which is suggestive of lichen planopilaris.

Skin biopsy was taken from abdominal lesion showed perifollicular lymphocytic infiltrate with damage to follicular basal cells which is suggestive of lichen planopilaris (Figure 4).

### 3. Discussion

We report a pediatric case of E/beta thalassemia with GLPLS which has never been described before. This dermatosis which is a variant of lichen planus may have a human leukocyte antigen (HLA) induced T cell response behind its clinical expression (7). Rodríguez Bayona *et al.* reported autoantibodies against Inner Centromere Protein INCENP, a protein responsible for chromosomal segregation and mitosis regulation, in one patient with GLPLS (8).

Though typically sporadic and nonfamilial, one case of familial pattern has been described (7), an association with hepatitis B vaccination (9) and a phenotypically female (genetically XY) patient with androgen insensitivity syndrome (testicular feminization) have also been reported (10). Our patient, however, did not receive any hepatitis B vaccination in the past. We performed abdominal ultrasound to confirm the female phenotype but karyotyping was not done as we lost the

patient to follow up.

In early lesions of lichen planopilaris there is perifollicular lymphocytic infiltrate at the level of isthmus and infundibulum, along with vacuolar changes of the outer root sheath. Progression of disease shows perifollicular fibrosis and epithelial atrophy at the level of isthmus and infundibulum which gives rise to a characteristic hourglass configuration. In advanced cases there is alopecia with vertically oriented elastic fibers that replace the destroyed hair follicles. Similar findings were seen in our case. End stage scarring alopecia without any visible hair follicle is called pseudopelade of Brocq (11).

Topical and systemic glucocorticoids, retinoids, and PUVA (psoralen and ultraviolet A) photochemotherapy have not shown to halt progression of the disease. Treatment with cyclosporine (4 mg/kg/day) has shown improvement in erythema, hyperkeratotic papules, and induction of partial hair growth in previous reports (12,13). In some cases improvement with thalidomide have also been described (14,15).

#### 4. Conclusion

The main aim of reporting this rare case is to make the clinicians aware of the occurrence of this rare syndrome in pediatric age group. If a patient presented with progressive cicatricial scalp alopecia associated with non-cicatricial alopecia of axillary and pubic areas and a follicular lichenoid eruption on the trunk and extremities then clinician should suspect GLPLS.

#### References

- Graham Little EG. Folliculitis decalvans et atrophicans. *Br J Dermatol.* 1915; 27:1835.
- Varadraj V Pai, Naveen N Kikkeri, Tukaram Sori, US Dinesh. Graham little piccardi lassueur syndrome: an unusual variant of follicular lichen planus. *Int J Trichology.* 2011; 3:28-30.
- Vashi N, Newlove T, Chu J, Patel R, Stein J. Graham Little Piccardi Lassueur syndrome. *Dermatol Online J.* 2011; 17:30.
- Zegarska B, Kallas D, Schwartz RA, Czajkowski R, Uchanska G, Placek W. Graham Little syndrome. *Acta Dermatovenerol Alp Panonica Adriat.* 2010; 19:39-42.
- László FG. Graham Little Piccardi Lassueur syndrome: case report and review of the syndrome in men. *Int J Dermatol.* 2014; 53:1019-1022.
- Somani N, Bergfeld WF. Cicatricial alopecia: classification and histopathology. *Dermatol Ther.* 2008; 21:221-237.
- Viglizzo G, Verrini A, Rongioletti F. Familial Lassueur Graham Little Piccardi syndrome. *Dermatology.* 2004; 208:142-144.
- Rodríguez-Bayona B, Ruchaud S, Rodríguez C, Linares M, Astola A, Ortiz M, Earnshaw WC, Valdivia MM. Autoantibodies against the chromosomal passenger protein INCENP found in a patient with Graham Little Piccardi Lassueur syndrome. *J Autoimmune Dis.* 2007; 4:1.
- Rebora A, Rongioletti F, Drago F, Parodi. Lichen planus as a side effect of HBV vaccination. *Dermatology.* 1999; 198:1-2.
- Vega Gutiérrez J, Miranda-Romero A, Pérez Milán F, Martínez García G. Graham Little Piccardi Lassueur syndrome associated with androgen insensitivity syndrome (testicular feminization). *J Eur Acad Dermatol Venereol.* 2004; 18:463-466.
- Mobini N, Loussaint S, Kamino H. Non-infectious erythematous papular and squamous disease. In: *Lever's Histopathology of skin.* (Elder DE, Elenitsas R, Johnson BL, Murthy GF, editors). 9th ed., Philadelphia: Lippincott William Wilkins, 2005; pp. 179-214.
- Bottoni U, Innocenzi D, Carlesimo M. Treatment of Piccardi Lassueur Graham Little syndrome with cyclosporin A. *Eur J Dermatol.* 1995; 5:216-219.
- Mirmirani P, Willey A, Price VH. Short course of oral cyclosporine in lichen planopilaris. *J Am Acad Dermatol.* 2003; 49:667-671.
- George SJ, Hsu S. Lichen planopilaris treated with thalidomide. *J Am Acad Dermatol.* 2001; 45:965.
- Boyd AS, King LE Jr. Thalidomide induced remission of lichen planopilaris. *J Am Acad Dermatol.* 2002; 47:967-968.

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